Single Hole High Flow Arteriovenous Fistula
A Characteristic Presentation of Rendu-Osler–Weber Disease in a Young Adult Treated by Endovascular Approach Case Report

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Summary

A high flow pial arteriovenous fistula in a twenty-years-old girl is described. The arteriovenous communication corresponds to a single hole fistula on the right rolandic area, with a giant venous ectasia. The patient presented seizures and left hemiparesis as symptoms. The fistula was embolized with glue obtaining total occlusion of the shunt. Hypotension was induced and valsalva manoeuver was done during the glue injection to reduce the flow into the fistula,however 26 hours after the procedure the patient bled resulting in a fatal outcome. The purpose of this paper is discuss the presentation of Rendu-Osler-Weber (ROW) in children and the therapeutic guidelines.

Introduction

Cortical high flow arteriovenous fistula constitutes the most common intracranial presentation of ROW disease in children. They may be the only manifestation of the disease, since epistaxis and telangiectasia are unusual in early life. ROW disease is a rare autossomal disorder with strong penetrance but variable expression, the diagnosis being based on the family history most of the time [1,2,7]. The brain arteriovenous malformation in children can be single or multiple, with angioarchitectural characteristics of nidus type or direct fistula [3].

Case Report

A twenty-year-old girl suffered two episodes of generalized seizure. The first occurred during pregnancy, in the seventh month and the second two months after delivery. Familial history of recurrent epistaxis was related by un uncle. After the second episode of seizure, clinical and imaging investigation was done. A cranial computed tomography (CT) scan revealed a mildly high density round lesion in the right Sylvian fissure with homogeneous enhancement by contrast medium (figure 1). The lesion produced mass effect, compressing the lateral ventricle and sulcus in the right cerebral hemisphere. T2- weighted magnetic resonance image showed a flow void area in the same region, corresponding to the huge ectatic vein in the Sylvian fissure (figure 2). Right carotid angiogram revealed a single hole high flow arteriovenous fistula on the rolandic area, supplied by the right angular artery draining to the superior sagital sinus through an insular vein. There was a giant venous ectasia immediately after the fistulous point (figure 3). The branches of the anterior and middle cerebral group were opacified by leptomeningeal anastomosis as well as through anterior and posterior communicating arteries. The embolization was done under general anesthesia with hypotension induced achieving a rate of 40 mmHg in the medium arterial blood pressure during the glue

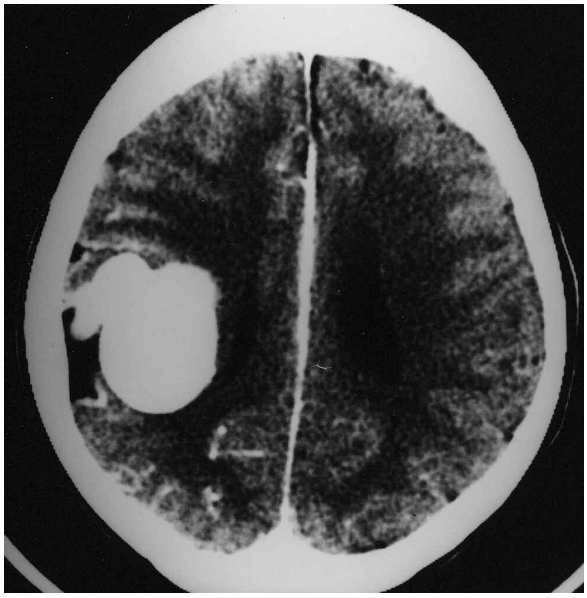

Figure 1 CT with contrast-Homogeneous enhancement by the contrast medium at the round lesion on the right rolandic area.

injection. Associated Valsalva maneuver was done to stop the diastolic flow and reduce the flow in the fistula, avoiding migration of the glue to the venous compartment. The transarterial approach was used, by the catetherization of the arterial feeder of the fistula, using a Sense-Nycomed microcatheter (figure 4). The embolic agent used was NBCA (Histoacryl) in a high concentration (98%) mixed with Tantalum powder. The control angiogram immediately after embolization showed subtotal occlusion of the shunt. The large amount of glue in the venous ectasia as well in the fistula itself prompted us to interrupt the procedure and to do a control angiogram in six months to confirm the total thrombosis of the shunt or otherwise to do another session of embolization to achieve complete occlusion of the fistula (figure 5). Clinically the patient improved with regression of the slight left hemiparesis presented on admission, and no further seizure episodes occurred. She did a control CT revealing that the fistula was not totally thrombosed, so a control angiogram was done six months after the first embolization confirming flow in the fistula and the cast of the histoacryl displaced slighted inside the venous pouch (figure 6). Another embolization without problems was done using the same embolic agent (Histoacryl) in a high concentration mixed with the Tantalum powder. Hypotension and Valsalva manoeuver were used as in the first embolization. The immediate control angiogram showed total occlusion of the fistula with redistribution of the blood flow to the middle and anterior cerebral group of arteries ipsilateral to the fistula (figure 7). Stasis inside the giant venous pouch indicating a progressive thrombosis as well at the venular compartment in the vicinity of the fistula was noted (figure 8). The patient was extubated and kept in Intensive Care Unit (ICU) under strict control of blood pressure with drugs (Nitroprusside). No hypertensive peak in the blood pressure was noted after the procedure. Four hours after the embolization she complained of headache followed by a decrease in consciousness level. She presented no clinical sign of haemorrhage. A CT was done that showed no bleeding. The symptoms regressed in the night, with a recovery of the consciousness level and no residual neurological deficit. A higher dose of Nitroprusside was used to achieve more reduction in the blood pressure level. On the next morning, twenty hours after the embolization, the patient presented a normal consciousness level, without any neurological sign or symptom, however no episode of variation in conciousness was remembered by the patient. Twenty-six hours after the embolization she complained of severe headache associated with vomiting followed by a rapid decrease in the conciousness level requiring intubation presenting a rapid evolution

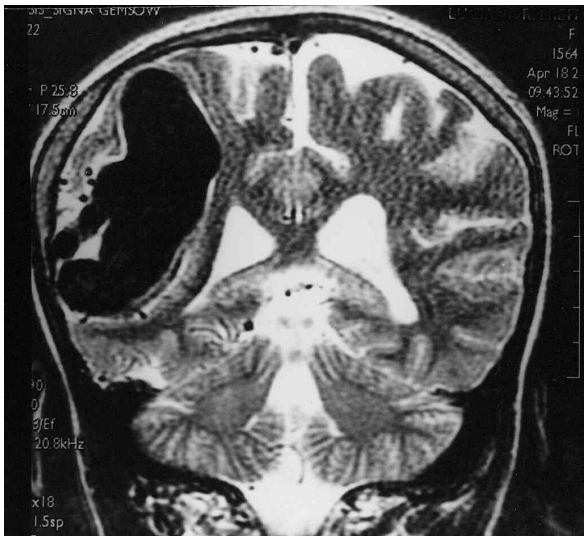

Figure 2 MRI T2W-Flow void area in the fistula point and the giant ectatic vein at the Sylvian fissure.

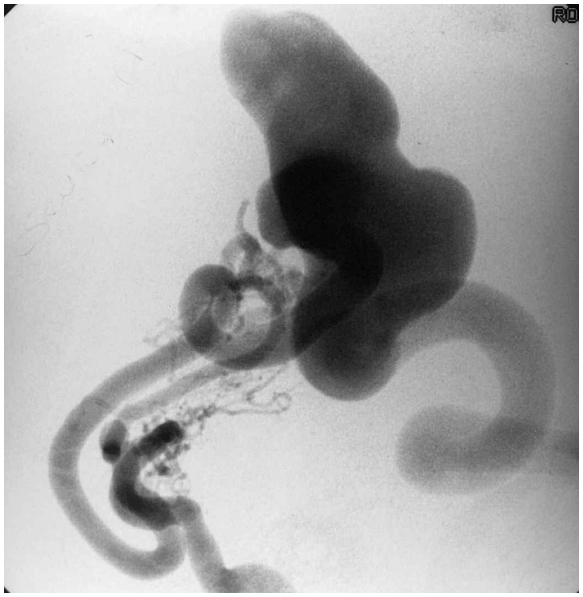

Figure 3 Right ICA angio. Note the single hole A-V fistula draining to a single cortical vein with a giant venous ectasia at the insular vein.

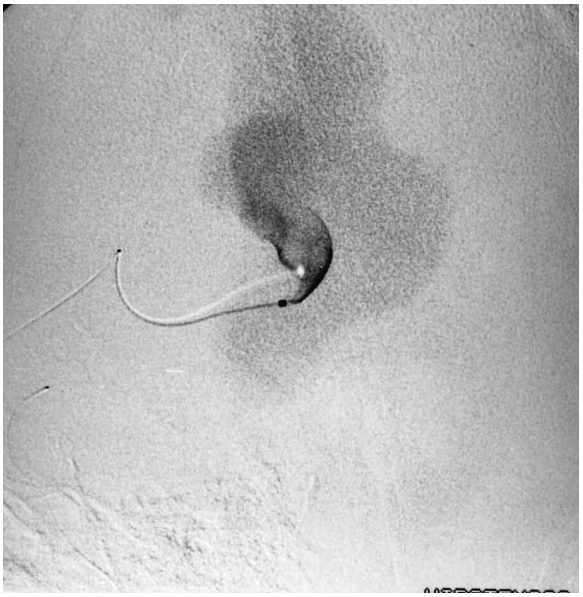

Figure 4 Superselective catheterization of the fistula. Note the position of the microcatheter against the wall of the artery.

to a coma state. At that time she presented a bilateral Babinski sign, right anisocoria (ipsilateral), fixed midriasis and decerebration (Glasgow scale-04). An urgent CT confirmed a huge right temporoparietal haematoma associated with herniation and mass effect (figure 9). Surgical treatment was done in emergency with decompressive lobectomy and haematoma evacuation. The surgical aspect revealed a diffuse profuse parenchymatous bleeding from the vicinity of the fistula, with major venous congestion. Major brain edema was present and it was difficult to stop the bleeding requiring brain resection. The patient never recovered and finally died.

Discussion

ROW disease is a rare autosomal dominant disorder characterized by multiple mucocutaneous and visceral telangiectasias which give rise to haemorrhagic complications in adulthood [1]. The pathologic process is a mutation of the endoglin receptor complex on endothelial cells. The endothelial cells lacking endoglin respond poorly to TGF-β creating abnormal vessels [3]. The TGEF-β is involved in controlling the remodelling process of the venules. There must be local conditions involved to transform a generalized genetic abnormality into a specific phenotype. The high flow fistula in infants occurs when the revealing trigger in the brain is effective either during a very limited window time or during a specific endothelial cell cycle [3]. So pial arteriovenous fistula reflects the fact that the morphology is precise in comparison with the date of its appearance in the patient's history [3]. This could explain why a new arteriovenous fistula never developed after the discovery of a given one, unlike facial or mucosal angioectasia [2,3]. In all reports, the BAVM or

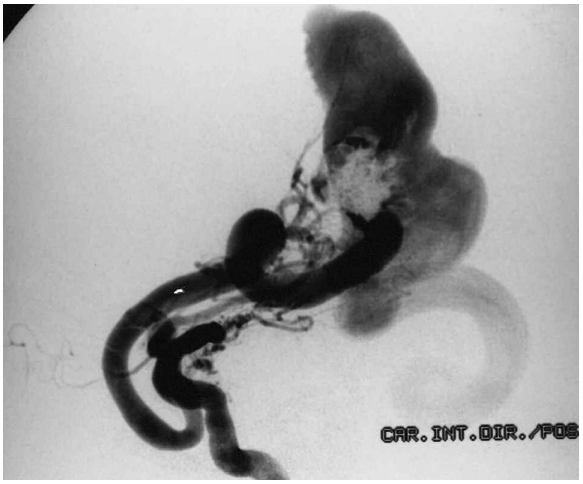

Figure 5 Control angiogram post embolization with subtotal occlusion of the shunt.

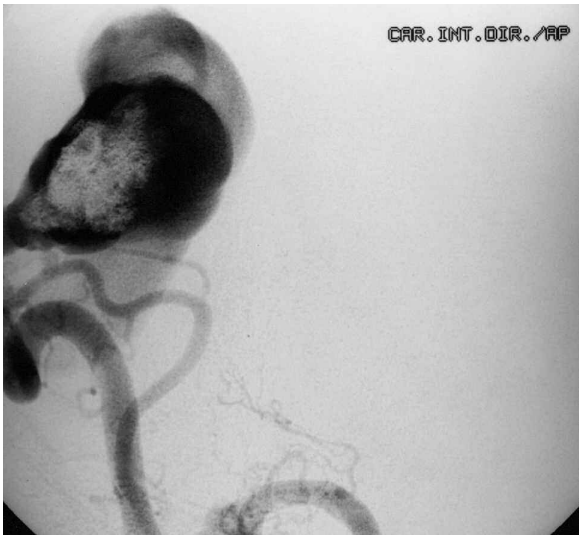

Figure 6 Control angiogram 06 months after the first embolization. Note the slight displacement of the glue inside the venous pouch.

AVF has a cortical location [1-3]. Arteriovenous fistulas are frequent in children and are always superficial and cortical. These shunts can be the sole manifestation of the disease in children, and are most of the time diagnosed in the first years of life [1,3]. Naturally, an organ where there is little turnover of cells is less likely to express a genetic disease. The typical symptoms of ROW are unusual before puberty because telangiectasias are not congenital, but acquired lesions, which appear to result from hyperactive angiogenetic factors, resulting in a focal dilatation of post capillary venules, that become tortuous associated with the excessive development of layers of smooth muscle without elastic tissue. The telangiectasias and haemorrhagic complications progress with age [1].

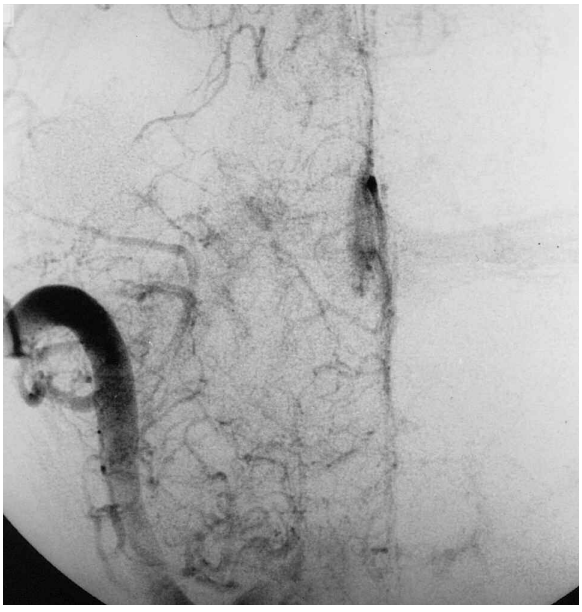

Figure 7 Immediate control angiogram post embolization-Late capillary phase. Note the total occlusion of the fistula and anterograde opacification of MCA and ACA territories. Note the early opacification of the deep venous system.

The central nervous system (CNS) AVM resemble the usual congenital types and should thus be distinguished from the telangiectasias. This suggests that they represent either an associated lesion or the embryonic expression of the capillary remodeling disorder of ROW. Whatever the etiology and pathogenesis of AVMs of the CNS in ROW, these lesions may be the presenting manifestation of the disease in children [1].

A high incidence of multiple CNS arteriovenous shunts has been described [1,4-6]. Willinsky et Al reported that 28% of patients with multiple CNS AVM had ROW, while associated AVM of CNS are present in approximately 8% of patients with ROW [4].

According to the literature, the difference between the AVF and AVM in patients with ROW is that multiplicity is more common in the AVM type, and the bleeding rate is higher in this group. The group of patients with AVM are more frequently symptomatic than the AVF ones [2]. Lasjaunias mentioned that AVF in children tent to harbour a high flow shunt which always plays an important role as an intraluminal stress trigger [3,16]. If so, the angioarchitecture of AVF in younger patients may have some specific features to induce intracranial bleeding. More than 80% of the cases had a direct cortical drainer with venous ectasia and almost all AVF had only one cortical feeder draining to a cortical vein [2]. Except for the high incidence of multiple lesions, pial AVF in children with ROW were identical to sporadic (non ROW children) pial AVF [1,2]. The clinical findings in children with pial AVF and ROW do not differ from those of any pial AVF [4,7,8]. Neurological deficits were probably caused by cerebral venous congestion and ischemia [1].The history, MR, CT and angiographic findings help to determine which lesion is causing the symptoms and needs prompt treatment. The clinical manifestations of pial AVF have a strict corre-

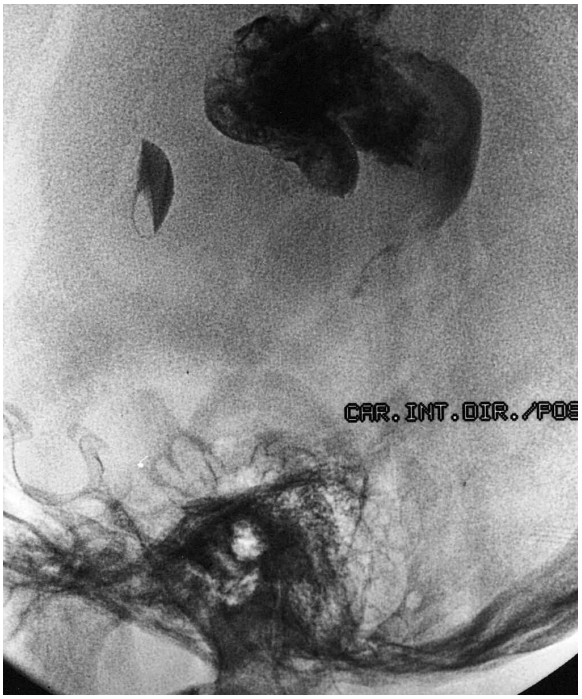

Figure 8 Note the cast of glue at the fistula site as well as inside the venous pouch. with stasis in the last one.

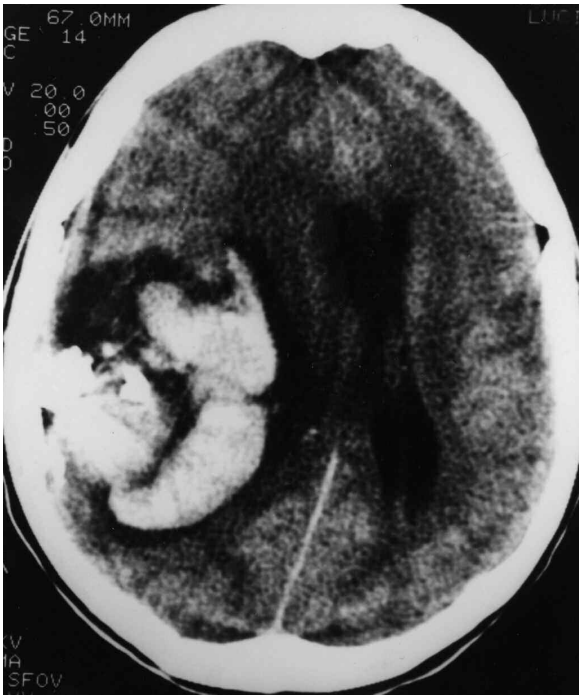

Figure 9 CT done 26 hours after embolization reveals a huge right hematoma with a severe mass effect.

lation with the age of the patient of the time of onset [9,10]. Cerebral bleeding is the most common manifestation of pial AVF in children [9,10,11].

The coexistence of BAVM and SCAVM is extremely rare [12-14]. Hasegawa reported only eight cases from the literature [13,14] of association of these two entities. However, as far as multiple BAVM are concerned, Iizuka [7] reported in the pediatric population that 20% of patients had multiple AVMs. In this situation, ROW shunt must be suspected. Furthermore, multiple lesions seen in ROW involve various metameric territories [15]. Treatment of asymptomatic pial AVF is controversial [9,10].

Lownie et Al [9] suggest observation of asymptomatic patients, but others workers [9,11] suggest active treatment to close the AVF, because of the poorer prognosis of conservatively managed patients. These patients have a high mortality of untreated cases - 05 deaths in 08 patients treated conservatively [11]. So total and definitive occlusion of a pial AVF is recommended whenever possible. Embolization of pial AVF is best accomplished by transarterial access. NBCA, detachable balloon or coils have been used. Our previous experience with CNS endovascular therapy led us to choose NBCA.

In the present case, the angiographic aspect of the high flow fistula with a giant venous ectasia in an adolescent patient, associated with a familial history of recurrent epistaxis induced us to diagnose ROW disease. The management was decided after a multidisciplinary discussion and endovascular treatment was done using an arterial approach.

The technical aspects undertaken during the glue injection (hypotension and valsalva manoeuver) were crucial to the occlusion of the fistula, avoiding the migration of the Histoacryl beyond the fistulous point.

The explanation of the bleeding post embolization prompted us to discuss several angioarchitectural aspects of the AVF.

The pathophysiologic mechanisms producing hemorrhage in a BAVM are related to arterial or anterograde pressure, venous or retrograde pressure or to hemorrhage secondary to ischemic infarction [17]. The concept of venous ischemia, accepted by most authors, is never recognized as the dominant one. Spetzler and Selman [18] commenting on Nornes and Grip's work [19], emphasized the role played by locally increased venous pressure, but only as a secondary or additional factor in decreasing tissue

perfusion. Like most other authors, they related ischemic symptoms mainly to the arterial steal. Even when discussing the "sump effect" after ressection of the AVM, only the capillary bed was considered [19,20,21,22], and its loss of autoregulation was proposed to explain the breakthrough perfusion theory first described by Spetzler [20]. Concerning the vein compartment, 60%-80% of the cerebral blood volume is located in the cerebral venules, where the flow is bidirectional in the white matter, allowing them to fulfill their nutrient role even in a retrogarde fashion [17].The congestion of the system that occurs either hemodynamically or by stasis, due to stenosis or thrombosis, will produce ischemic phenomena of venous origin and accumulation of catabolic substances in the parenchyma. At some point, the white matter can no longer tolerate the venous congestion. The acquired nature of the congestion quickly makes it become symptomatic. The progressive nature of the deficits and their fluctuation in severity reflect the attempts of the collateral circulation to overcome the increased pressure in the venous outlet of the shunt. Decreased tissue perfusion secondary to venous ischemia may produce virtually any type of neurological symptom such as motor or sensory deficits, neuropsychological alteration and seizures [17].

In the present case, we believe that the ischemic phenomenon of venous origin was created by the total occlusion of the fistula with thrombosis of the giant venous pouch leading to venular dysfunction of the tissue surrounding the fistula region secondary to hyperpressure resulting in a fatal outcome.

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